Disorders of Esophageal Motility

These discussions are selected from the weekly staff conferences in the Department of Medicine, University of California Medical Center, San Francisco. Taken from transcriptions, they are prepared by Drs. Martin J. Cline and Hibbard E. Williams, Assistant Professors of Medicine, under the direction of Dr. Lloyd H. Smith, Jr., Professor of Medicine and Chairman of the Department of Medicine.

DR. SMITH*: The patient this morning will be presented by his private physician, Dr. Howard Shapiro.

DR. SHAPIRO*[1]: Thank you, Dr. Smith. The patient is a 28-year-old Caucasian male who had had known difficulty in swallowing since childhood. This dysphagia was more pronounced with solids than with liquids, and he would frequently regurgitate undigested food eight to ten hours after a meal. In the several years before admission to hospital, his difficulties were complicated by substernal discomfort which generally occurred at night and would awaken him from sleep. This discomfort was relieved somewhat by antacids. He had no difficulty in maintaining his nutrition. A trial of antacid therapy and of viscous xylocaine failed to relieve the symptoms of dysphagia.

Results of physical examination were within normal limits. An upper gastrointestinal series revealed cardiospasm with moderate dilatation of the upper esophagus. Early vermiform movements and secondary peristaltic stripping motions were demonstrated at fluoroscopy. Esophageal motility studies showed a normal appearing gastroesophageal sphincter. There was no relaxation of the gastroesophageal sphincter to swallowing, and the swallowing waves were simultaneous throughout the esophagus. There was no evidence of peristalsis on the motility studies.

*Lloyd H. Smith, Jr., M.D., Professor & Chairman, Department of Medicine.

*[1]Howard A. Shapiro, M.D., Assistant Clinical Professor of Medicine.

An esophagogastric myomectomy (Heller procedure) was done through a transverse abdominal incision. Since the time of operation the patient has remained quite well with the exception of some heartburn early in the recovery period. Follow-up esophageal motility studies continued to show a failure of peristalsis with simultaneous appearance of the swallowing waves throughout the length of the esophagus.

(The patient enters.)

DR. SHAPIRO: Could you describe the major symptoms that you had before coming to my office?

PATIENT: I could not swallow food very well and I regurgitated two to eight hours after eating. It was rather painful at times.

DR. SHAPIRO: How much weight have you gained since the operation?

PATIENT: I weigh about five pounds more than I did before the operation.

DR. SHAPIRO: Have you had any symptoms since?

PATIENT: Not really.

DR. SHAPIRO: Have you noticed any heartburn since the operation?

PATIENT: No.

DR. SMITH: Dr. Dietschy, would you like to bring out any points?

DR. DIETSCHY*[2]: I wonder if you would comment in more detail on the pain which you had at the very beginning of your troubles. Were you having chest discomfort at any time of the day or night?

PATIENT: No, the pain did not start until about two and a half years ago, and I had trouble swallowing for about five years before that.

DR. DIETSCHY: You were a teenager when this began?

PATIENT: Yes.

DR. DIETSCHY: Since then, you are sure that you have had no burning sensation in your chest?

PATIENT: It feels more like gas.

DR. DIETSCHY: Is it a sense of fullness rather than a burning sensation?

PATIENT: Yes.

DR. DIETSCHY: Does it tend to rise up in your chest?

PATIENT: Yes.

DR. DIETSCHY: And is this relieved by taking antacids?

PATIENT: No, it is not.

DR. DIETSCHY: Thank you. (Patient leaves.)

DR. SMITH: The x-ray films will be discussed by Dr. Zboralske.

DR. ZBORALSKE*[3]: A preoperative study demonstrated pronounced dilatation of the esophagus. The lower esophagus reveals a typical bird-beaking appearance. No peristalsis was identified during the time of fluoroscopy. Postoperative studies demonstrated much less dilatation of the esophagus but some persistent mild narrowing distally, although it does empty quite promptly. Again, no esophageal peristalsis was observed at fluoroscopy.

DR. SMITH: We are delighted to have as our major discussor this morning, Dr. John M. Dietschy from the University of Texas Southwestern Medical Center in Dallas. He is a member of the Department of Medicine there and assistant professor of medicine at the University of Texas Southwestern Medical School. Today he will discuss disorders of esophageal motility. Dr. Dietschy.

*[2]John M. Dietschy, M.D., Assistant Professor of Medicine, University of Texas Southwestern Medical School.

*[3]F. Frank Zboralske, M.D., Assistant Professor of Radiology.

Disorders of Esophageal Motility

DR. DIETSCHY: The patient presented by Dr. Shapiro manifests many of the findings commonly seen in the esophageal disorder known as achalasia. I would like to use the problem presented by this patient as the basis for a brief review of the normal physiology of the esophagus, and then to discuss two disorders of esophageal motility: Tertiary contractions and achalasia.

The esophagus is a muscular tube, made up of an outer longitudinal and an inner circular layer of muscle, which connects the lower end of the pharynx with the stomach. It is closed off both at its upper and at its lower ends by barrier mechanisms which prevent the inadvertent entrance of secretions and food from the mouth and stomach into the body of the esophagus. Anatomically the superior barrier mechanism consists primarily of a specialized bundle of the inferior pharyngeal sphincter, the cricopharyngeous muscle, which inserts on the lateral margins of the cricoid cartilage and passes strap-like around the posterior aspect of the esophagus, where it can act as an efficient sphincter.

The inferior barrier mechanism, on the other hand, is much more complicated and controversial. There is no specialized muscle band demonstrable at the lower end of the esophagus in the human, although such an anatomic structure can be found in certain lower animals. Nevertheless, there is a discrete area found at the lower end of the esophagus, the so-called high pressure zone, which behaves physiologically as a sphincter. Other mechanisms which are thought to contribute to the inferior barrier include the diaphragmatic sling formed around the esophagus as it passes through the diaphragmatic hiatus, the acute angle present at the esophagogastric junction, rosette formation by gastric mucosa pushing up into the base of the esophagus, and finally, the intra-abdominal portion of the esophagus which is subject to intra-abdominal pressure. While all of these may contribute together to the inferior barrier mechanism, of particular importance to the discussion today is the high pressure zone or inferior esophageal sphincter.

The Sequence of Events in Swallowing

The sequence of events which occur in the esophagus during the normal act of swallowing may be followed by placing pressure-sensing devices

at various levels of the pharynx, esophagus and stomach. In the resting esophagus two areas of elevated intraluminal pressure can be demonstrated; these are located at the levels of the upper and lower esophageal sphincters. Within the body of the esophagus one finds a slightly negative pressure with variations in the baseline caused by the respiratory cycles. On the abdominal side of the diaphragmatic hiatus, the respiratory variations are reversed, and the pressure in this area is usually close to atmospheric pressure.

Swallowing begins as the tongue is pushed up against the hard palate and swept backwards as the nasal pharynx is closed and the pharyngeal sphincters contract. This results in the development of an abrupt rise in pressure in the hypopharynx. Just preceding these events in the hypopharynx, however, the superior esophageal sphincter relaxes, allowing the liquid or solid bolus to be pushed into the upper portion of the esophagus by the pharyngeal pressure wave. Here there is a momentary delay following which a primary peristaltic wave is initiated in the upper part of the esophagus. This peristaltic wave is then propagated down the body of the esophagus at a rate of 2 to 3 cm per second. Just before the arrival of the primary peristaltic wave at the lower esophageal sphincter the resting pressure in this area relaxes so that the bolus may pass through the esophagogastric junction into the stomach.

This sequence may be observed easily at fluoroscopy. As the patient begins to swallow and high pressure develops in the hypopharynx, a liquid bolus of barium suspension will be forced downward and almost instantaneously fill the entire body of the esophagus. In contrast, if the bolus is solid food, then the pharyngeal pressure wave merely pushes it into the top of the esophagus where it remains momentarily until primary peristalsis is initiated. In either case one then observes a contraction wave which begins at the upper end of the esophagus and moves downward pushing the bolus ahead of it. Just before the arrival of this wave at the lower end of the esophagus the lower sphincteric region opens and the bolus is conveyed into the cardiac portion of the stomach.

Primary peristalsis, then, is that integrated series of events seen in the esophagus which is initiated by the act of swallowing and whose function is to convey liquids and solids from the mouth into the stomach.

Secondary Esophageal Peristalsis

A second type of normal motility is known as secondary peristalsis. In this situation, peristalsis is not initiated by the act of swallowing, but by distension of some part of the body of the esophagus. If the esophagus is distended at a particular level, then at this point a peristaltic wave is initiated; the wave sweeps down the esophagus and the lower esophageal sphincter relaxes as it does with a primary peristaltic wave. There is no retrograde conduction of peristalsis upward from the point of distension and no relaxation of the superior sphincter occurs.

One can observe this sequence occurring in secondary peristalsis under the fluoroscope. For example, in a patient with esophageal regurgitation, one may observe reflux of barium meal from the stomach back into the body of the esophagus. The barium seldom moves upward very far, however, before the esophagus responds with a secondary peristaltic wave which sweeps downward, clearing the esophageal lumen of the regurgitated barium.

Secondary peristalsis, then, is that integrated series of events seen in the esophagus which is initiated by distension of the body of the esophagus and whose function is to clear the esophageal lumen of material refluxed from the stomach or left behind in the body of the esophagus by the primary peristaltic wave.

Abnormal Motility

I would like now to discuss two forms of abnormal esophageal motility which give rise to distinct clinical syndromes. The first of these is the syndrome of tertiary contractions in which the motility disorder involves primarily the lower half of the body of the esophagus but spares the lower esophageal sphincter; the second is achalasia (cardiospasm) in which abnormal function of the lower half of the esophagus as well as the inferior esophageal sphincter can be demonstrated.

In tertiary contractions normal pharyngeal and superior sphincteric mechanisms are present, but instead of a primary peristaltic wave sweeping down the esophagus in an orderly, propagated manner, there is disordered, irregular contraction of the entire lower one-half or two-thirds of the body of the esophagus with the development of very high intraluminal pressures. Furthermore, in response to a single swallow one may observe repetitive contractions of this area of the esophagus. Yet, despite this abnormality of peristaltic

activity in the body of the esophagus, the lower esophageal sphincter does relax; this is an extremely important point in differentiating this syndrome of tertiary contractions from the syndrome of achalasia.

Tertiary contractions appear quite dramatic when viewed at fluoroscopy. They may be mild, appearing only as indentations or slight rippling motions in the body of the esophagus. Not infrequently they are more severe with irregular contractions of the entire length of the esophagus, or in some patients one may occasionally observe pronounced segmental contractions with pseudodiverticulum formation. This lesion has made a tremendous impression on the imagination of roentgenologists, who have applied many colorful and confusing terms to this lesion. Schatzki was the first to refer to this motility as "Kräuselung"—that is, rippling or curling of the esophagus. Subsequently, this motility disorder as seen roentgenographically has been referred to as the rosary-bead deformity, the string-of-pearls deformity, the corkscrew esophagus, spastic pseudodiverticulosis or segmental diverticulosis, and, finally, as the Barony-Teschendorf syndrome—a syndrome named for two German roentgenologists who first described this disorder. There is neither experimental nor clinical evidence to separate any of these particular forms of the motility disorder. I believe, therefore, that these should all simply be referred to as "tertiary contraction" of the esophagus.

Characteristics of Tertiary Contractions

This disorder of esophageal motility is commonly seen during examination of the upper gastrointestinal tract, particularly in patients over the age of 50. Attacks of tertiary contractions are commonly transient, and between episodes patients are usually found to have entirely normal primary and secondary peristaltic activity. Fortunately, the vast majority of patients found to have tertiary contractions have no symptoms related to this motility disorder. Very rarely, however, dysphagia and substernal pain associated with tertiary contractions of the esophagus develops. It is this clinical syndrome which is known as "symptomatic tertiary contractions" or "diffuse spasm" of the esophagus. In general there is no correlation between the apparent severity of the tertiary contractions as seen fluoroscopically and the severity of the pain-dysphagia syndrome associated with these attacks.

The pain of symptomatic tertiary contractions is fairly characteristic. It is typically substernal in location and in severe cases may radiate into the neck or shoulders. It varies in quality from a rather dull, uncomfortable sensation to a severe, constricting pain beneath the breast bone. It can mimic the pain of myocardial infarction in every way and, furthermore, may be relieved by nitroglycerin and amyl nitrate. Attacks of symptomatic tertiary contractions are usually initiated by those events which normally activate primary or secondary peristaltic activity in the body of the esophagus. Thus, the act of swallowing may precipitate an attack of pain and dysphagia. Similarly, the recumbent position, which enhances gastro-esophageal reflux, may initiate episodes of substernal pain in the middle of the night. Such attacks are quite irregular in frequency, duration and intensity. Since the patient may manifest entirely normal esophageal motility between attacks, the diagnosis depends upon the ability to reproduce the syndrome under fluoroscopic control when the pain and dysphagia can be shown to occur coincident with tertiary contractions of the body of the esophagus.

Since the inferior sphincter relaxes normally in this syndrome, there is usually little or no dilatation of the body of the esophagus. In addition, the esophagus in this condition does not manifest hypersensitivity to mecholyl—that is, the mecholyl test result is negative.

Achalasia

In the syndrome of achalasia, there is also disordered motility of the lower one-half to two-thirds of the body of the esophagus; but, equally important, there is also dysfunction of the lower esophageal sphincter in that this area of musculature fails to relax in response to motor activity in the body of the esophagus. Consequently, there is progressive dilatation and elongation of the esophagus in this disease. Early in the syndrome one commonly sees irregular tertiary contractions of the body of the esophagus, but as dilatation progresses and the musculature becomes atonic, these irregular contraction waves tend to disappear. The motility disorder in achalasia, then, is twofold: (1) There is loss of normal primary and secondary peristaltic activity in the lower portion of the body of the esophagus; and (2) the inferior esophageal sphincter fails to relax properly during the act of swallowing, and this functional obstruction, in turn, leads to progressive dilatation.

Patients with achalasia may manifest both substernal pain and dysphagia, but these symptoms usually begin at an earlier age than those associated with tertiary contractions. The initial symptoms of dysphagia may be irregular in intensity and occur at only irregular intervals. Eventually, however, difficulty in swallowing becomes progressively worse leading to weight loss. There may also be a sensation of substernal fullness and episodes of choking, particularly at night, when esophageal contents spill into the bronchopulmonary tree.

The diagnosis of this syndrome rests on the fluoroscopic demonstration of lack of primary and secondary peristaltic activity in the lower half of the esophagus and failure of the lower esophageal sphincter to relax properly. In addition, the body of the esophagus in achalasia responds to small doses of mecholyl with an exaggerated tetanic contraction (positive reaction to a mecholyl test).

Thus, these two disorders of esophageal motility may be summarized as follows:

Symptomatic tertiary contractions are primarily seen in elderly patients, whereas in achalasia symptoms classically begin in early middle age.

In achalasia there is motor dysfunction both of the body of the esophagus and of the inferior esophageal sphincter. In tertiary contractions, in contrast, the motility disorder appears to be limited to the body of the esophagus.

A positive reaction to a mecholyl test is seen in patients with achalasia but usually not in those with symptomatic tertiary contractions.

Moderate to pronounced dilatation of the esophagus is common in achalasia but is usually not seen in symptomatic tertiary contractions.